CASE REPORT

Successful pregnancy and delivery in a woman with a single ventricle and Eisenmenger syndrome

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Background: **Patients with a single ventricle represent a rare abnormality found in 1% of patients with congenital heart disease, often discovered during childhood. Without pulmonary stenosis, the disease can progress to fixed pulmonary hypertension. Both pregnancy and delivery are risky events capable of increasing the right-to-left shunt. Pregnancy is contraindicated.**

Case: **We report the case of a 27-year-old woman with a single ventricle without pulmonary protection and fixed pulmonary hypertension at 60 mmHg, discovered during a pregnancy. The delivery was obtained by cesarean section with epidural anesthesia and the patient was perioperatively treated with nitric oxide. Though contraindicated, pregnancy and delivery were successfully achieved in this patient.**

Comment: **Patients with single ventricle and Eisenmenger syndrome rarely reach adult life. Pregnancy with this condition is exceptional and fundamentally perturbs hemodynamic stability. In spite of the development of anesthesia and resuscitation and the description of some cases in literature, pregnancy with Eisenmenger syndrome is contraindicated.**

Keywords: Pregnancy, Single ventricle, Eisenmenger syndrome

Introduction

Eisenmenger syndrome is defined as a pulmonary vascular obstructive disease developing as a consequence of a large pre-existing left-to-right shunt such that pulmonary artery pressures approach systemic levels and the flow becomes bidirectional or right-to-left [1].

The occurrence of a single ventricle is rare, often discovered during childhood. Without pulmonary stenosis, the disease leads to Eisenmenger syndrome. Both pregnancy and delivery are risky events associated with high morbidity and mortality rates. The presence of pulmonary hypertension is considered to be one of the major maternal risk factors [2]. Therefore, successful pregnancy

Disclosure: Authors have nothing to disclose with regard to commercial support.

Received 14 March 2013; revised 10 April 2013; accepted 21 May 2013.
Available online 29 May 2013

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1016–7315 © 2013 Production and hosting by Elsevier B.V. on behalf of King Saud University.

Peer review under responsibility of King Saud University.
URL: www.ksu.edu.sa
http://dx.doi.org/10.1016/j.jsha.2013.05.001

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associated with this condition is rare. Nevertheless, some women prefer to run the risks despite medical advice.

Case

A 27-year-old woman (gravida 1, para 0) was admitted at 19 weeks gestation for dyspnea and palpitations. She had reported effort intolerance since childhood but had never been investigated. Physical examination showed clubbing of fingers and cyanotic lips. Her oxygen saturation by pulse oximetry (SpO_2) was 70%. Her vital signs were as follows: heart rate of 80 bpm, blood pressure of 100/60 mmHg and respiratory rate of 22 breaths per minute. On cardiac auscultation, a systolic murmur could be heard over the second left intercostal space with an accentuated single second heart sound. Laboratory analysis showed hemoglobin of 16.1 g/dl and hematocrit of 49%. No abnormality was found in her chest X-ray. An electrocardiogram showed a sinus rhythm with a right bundle branch block. An echocardiography revealed a double inlet single with left ventricular morphology and atrioventricular concordance. The right ventricle was rudimentary and the mixing was free between the two ventricular chambers. The great vessels were normally positioned (ventriculo-arterial concordance) with no evidence of pulmonary stenosis. The systolic performance of a single ventricle was conserved (ejection fraction = 60%) (Fig. 1). Also noted on the echocardiography was a moderate pulmonary and tricuspid regurgitation with a pulmonary hypertension of 60 mmHg (Fig. 2).

Figure 1. Double inlet single ventricle with conserved systolic performance (ejection fraction = 60%).

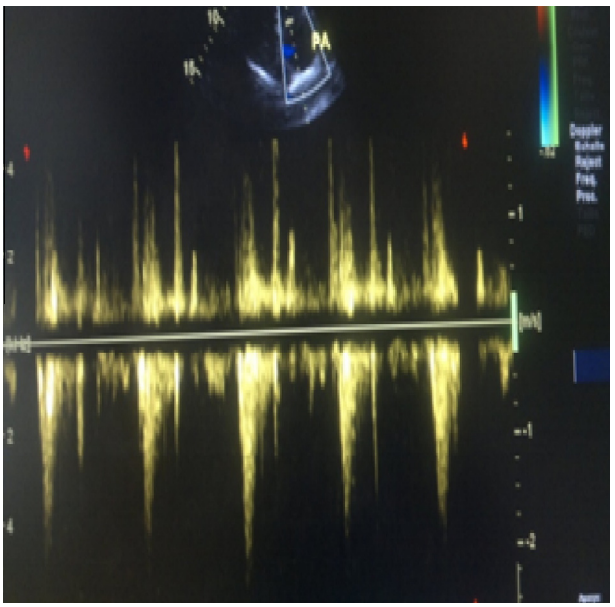

Figure 2. Moderate pulmonary regurgitation with a pulmonary hypertension of 60 mmHg.

A routine abdominal sonogram showed a normal living fetus. The diagnosis of a single ventricle with Eisenmenger syndrome was made and medical abortion was indicated but refused by the patient.

The woman was lost to follow up until she was admitted in labor at 37 weeks gestation. She was hemodynamically stable. A 22-gauge epidural catheter was inserted 3 cm into the epidural space. After injection of anaesthetic solution (bupivacaïne 5 mg and sufentanil 2.5 μg), the patient was placed in the supine position with left lateral tilt to avoid aortocaval compression. The delivery was obtained by cesarean section.

The infant was a male weighing 3000 g who had an Apgar rating of 7/9/10 after 1, 5 and 10 minutes. He did not show any malformations. During the first 48 hours, the patient received oxygen (6L/mn) in association with nitric oxide (initially 800 ppm for eight hours then 10 ppm) through a nasal cannula. The procedure was covered by antibiotics: an association of amoxicillin and clavulanic acid prescribed for five days. An anticoagulation therapy was done as follows: unfractionated heparin was introduced eight hours after the operation, and replaced by low molecular weight heparin 16 hours later before starting oral anticoagulants from the third day.

The post operative echocardiography noted the preservation of the left ventricular systolic function and the stability of pulmonary hypertension.

The echocardiography of the newborn was normal.

The post-partum period was uneventful, and the patient was discharged home on the thirteenth day. Three months later, inhibitors of endothelin receptors (Bosentan) were introduced. The mid-term outcome was good.

Comment

Single ventricle is a rare abnormality found in 1% of patients with congenital heart diseases [3]. Different forms can be observed; those without a pulmonary protection may lead to Eisenmenger syndrome [2]. Early diagnosis and banding of the pulmonary artery are able to prevent it. In our patient, it was too late and the pulmonary hypertension was fixed.

In such a condition, pregnancy makes dyspnea worse because of the increase of the cardiac output and the anemia it induces. Delivery represents a risky event too. In fact, it is responsible for the decrease of systemic vascular resistance and the increase of the pulmonary because of distal microthrombosis and hypoxia [3,4].

Severe complications such as heart failure, endocarditis and thromboembolic accidents can occur. They must be prevented by nitric oxide, antibiotics and anticoagulant therapy [4]. It has been demonstrated that in situ thrombi can develop and may worsen pulmonary hypertension. Therefore, anticoagulation, generally well tolerated, is recommended to prevent thrombus formation. The cessation of heparin administration during postpartum hemorrhage can be responsible for the rapid progression of pulmonary hypertension [5,6]. Inhaled nitric oxide is a selective pulmonary vasodilator for patients with pulmonary hypertension [7]. Endothelium-dependent relaxation of pulmonary arteries is impaired in Eisenmenger syndrome [8]. Direct inhalation of nitric oxide may reduce pulmonary hypertension and improve oxygenation by the optimization of ventilation-perfusion quotients. Inhaled nitric oxide also has an antithrombotic effect [9]. In Eisenmenger syndrome, it has been used not only in the management of labor but also as a bridge to heart-lung transplantation [10].

The mode of delivery is controversial. During labor, uterine contraction causes autotransfusion and may increase cardiac output by 25%. This increases pulmonary arterial pressure and may precipitate heart failure or arrhythmia. For these reasons and its shorter duration, cesarean section is commonly preferred to vaginal delivery [11].

Some authors do prefer general anesthesia over regional when anesthetizing a patient with Eisenmenger syndrome. In fact, regional anesthesia is potentially risky because it may decrease the systemic vascular resistance, which would increase the shunt and exacerbate hypoxemia [6]. Others recommend regional anesthesia with single ventricle [12,13]. Epidural anesthesia was successfully used in our case and provided excellent analgesia.

The prognosis of such cases depends on the pulmonary hypertension which represents the major mortality and morbidity factor [2].

The successful outcome of this pregnancy may be associated with the preserved systolic function of the single ventricle.

In spite of the development of anesthesia, resuscitation and the description of some cases in literature [14], pregnancy with Eisenmenger syndrome continues to be contraindicated because of a high dissuasive maternal mortality rate [6]. Each patient should be assessed by a combined and experimented obstetric, anesthetic and cardiology team.

Conclusion

Patients with single ventricle and Eisenmenger syndrome rarely reach adult life. Pregnancy with this condition is exceptional and fundamentally perturbs hemodynamic stability. Single ventricle performance and fixed pulmonary hypertension represent the most important prognostic factors. The mortality and morbidity rate is very high. Therefore contraception is essential and pregnancy is contraindicated.

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